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Scientific letter

Vanishing Lung Syndrome: Fifteen Years After Bullectomy

Síndrome del pulmón evanescente: quince años después de una bullectomía

Dear Editor:

The rare finding of large lung bullae associated to paraseptal and centroacinar emphysema described in young smokers has been coined as vanishing lung syndrome. This is a disorder without a clear cut etiology mainly reported in young tobacco, marijuana and hashish smokers. It is thought that prolonged breath holding and the toxic effects of the smoke leads to barotrauma and to the development of bullae.[1–4] The management of patients with vanishing lung syndrome may be complicated by the diagnostic confusion between lung bullae and pneumothorax.

We present a 37 year old smoker with a daily consumption of 10 joints (tobacco + hashish) during 20 years (200 joints-year) who complained of 1 year history of grade I dyspnea on the mMRC scale that had recently worsened. He did not improved with inhaled bronchodilators and the semiology was not suggestive of a respiratory infection. He was a well nourished man, body mass index of 24, in no respiratory distress. The chest auscultation revealed absent lung sounds on the right side. The radiograph is shown (Fig. 1, Image A). A recent spirometry and blood gas analysis were as follows: FEV_1: 1.93 L (46% pred.), FVC: 3.44 L (81% pred.), FEV1/FVC: 56%, PaO_2: 62 mm Hg, $PaCO_2$: 40 mm Hg. At that point, a complete

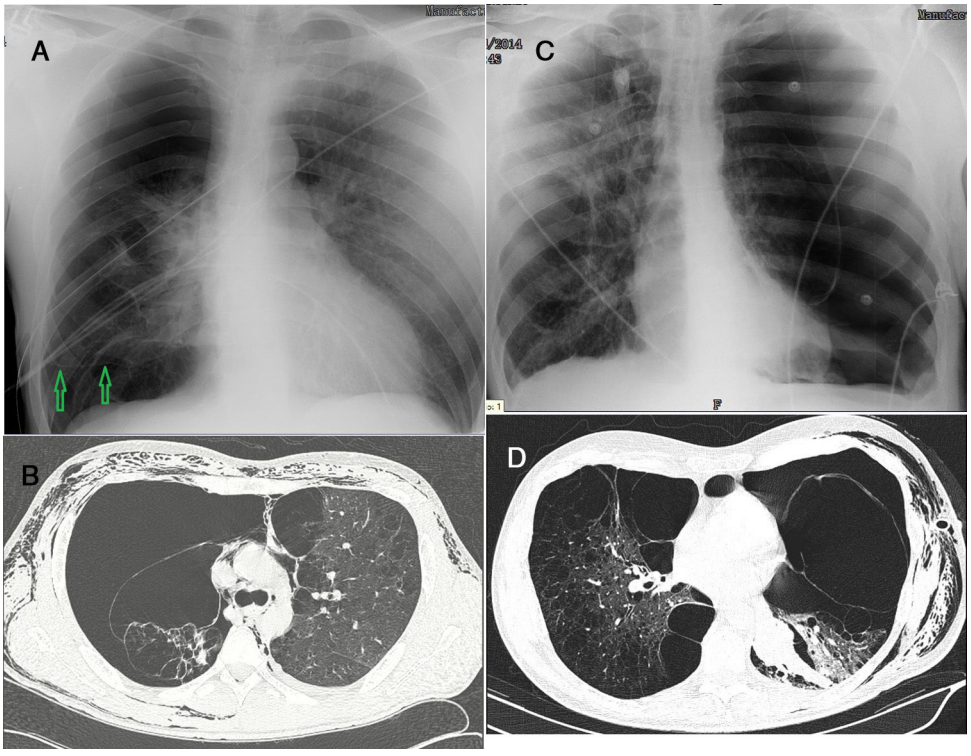

Fig. 1. (A) Preoperative chest radiograph. Right hemithorax hyperlucency with absent vascular markings in the upper field and partially in the lower field. A horizontal line corresponding to the visceral pleura is seen in the lower field (arrows) and appeared after the placement of a chest drainage. (B) CT image, axial view. Compressed right upper lobe bullae by iatrogenic pneumothorax and paraseptal and centroacinar emphysema in the left upper lobe. Mediastinal and subcutaneous emphysema are noted. (C) A complete left pneumothorax and a bulla with mediastinal shift toward the right side is seen. (D) CT image, axial view. Left pneumothorax, ipsilateral compressed upper lobe bulla and paraseptal and centroacinar emphysema in the remaining ipsilateral and contralateral lung. Subcutaneous emphysema and a left sided chest tube are seen. Atelectatic lung, presumably the left lower lobe, is visible.

pneumothorax was suspected and a chest tube was inserted without improvement in the patient's status. A representative image of the CT is shown (Fig. 1, Image B). The drainage was withdrawn and the patient underwent surgery with evidence of large bullae replacing the upper lobe and part of the lower lobe. A bullectomy and pleural abrasion were performed through video thoracoscopic assisted surgery (VATS), which eventually resulted in remarkable symptomatic improvement. The serum alpha 1 antitrypsin level was 140 mg/dl. He was advised to quit smoking and to follow up in the outpatient clinic. A follow up room air PaO_2 was 87 mm Hg, $PaCO_2$ was 40 mm Hg, and a repeated spirometry read FEV_1: 3.99 L (81% pred.), FVC: 6.70 L (134% pred.), FEV_1/FVC: 50%.

However, the patient was subsequently lost to follow up and presented 10 years later with increasing dyspnea. He admitted to ongoing smoking. The radiograph and a representative image of the CT scan are shown (Fig. 1, Images C and D). A new bullectomy and pleural abrasion were performed. A persistent air leak was a significant postsurgical complication and the patient was discharged home with a Heimlich valve. Once the valve was withdrawn, a follow-up spirometry read as follows: FEV_1: 1.83 L (48% pred.), FVC: 5.45 L (115% pred.), FEV_1/FVC: 33%. With such postoperative pulmonary function results and looking in retrospect, we acknowledge that it is doubtful whether the repeated bullectomy was appropriate given a likely low presurgical FEV_1.

After the second surgery the patient quit smoking and did not present significant exacerbations in the following 5 years. His pulmonary function remained somehow stable but clearly worse than after the first bullectomy. On a follow-up visit he walked 536 meters (predicted 630 meters) on a six minute walking test, his dyspnea on the Borg scale went from 0 to 5 and a fall in SpO_2 from 96% to 78% ensued. His current treatment includes a LABA/LAMA combination and he was offered a portable oxygen unit, but declined.

The diagnosis of vanishing lung syndrome and active smoking are considered a contraindication to bullectomy.[1] However, on the first occasion this patient showed criteria that predicted a good surgical outcome, such as young age, dyspnea despite medical therapy, FEV_1 higher than 40% predicted, normal $PaCO_2$, bullae occupying more than one third of the hemithorax with vascular crowding, and presumably healthy compressed pulmonary parenchyma. A pulmonary angiography or a perfusion lung scan was not done, so the latter statement remained hypothetical.

Bullous lung disease associated with marijuana use has long been observed in clinical practice but published evidence is limited to a total of about 65 cases.[5–15] Case reports tipically describe young patients with pneumothorax, pneumomediastinum and/or bullous lung disease – including the so called vanishing lung syndrome – who are marijuana and tobacco smokers. However, there is a need for epidemiological studies to confirm the apparent cause-effect relationship between marijuana or hashish smoking with bullous lung.[2] The patient we describe was a heavy smoker of tobacco plus hashish, which we think increased the risk.

A distinction between pneumothorax and lung bullae may be difficult on a plain radiograph. The presence of concave lines in relation to the chest wall suggest bullae as opposed to a convex line on upright films which usually corresponds to the visceral pleura in pneumothorax. In case of doubt a chest CT may aid in this distinction and help to avoid placing an unnecessary drainage.[15] After the first bullectomy, the finding of a high FVC despite airflow limitation is in agreement with what has been reported in some marijuana smokers.[2,3]

Given the high and increasing prevalence of marijuana and hashish smoking in our society, it is relevant to be aware of this rare pathology because of the substantial medical and financial impact that may have onto patients, some of whom might become lung transplant candidates.[3,5,7]

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